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## ABSTRACT

**Aim.** The aim of this study was to explore the meaning of living with uncertainty for people diagnosed with motor neurone disease (MND).

**Background.** MND is a progressive neurodegenerative condition resulting in multiple needs, arising from the complex nature of the disease trajectory. People with MND are often required to make decisions for symptom management and end-of-life care. Research into the lived experience of MND has previously highlighted: the shock of receiving such a diagnosis and prognosis; subsequent concerns relating to the future and loss; and the existential suffering for a person with MND. The lived experiences of MND accentuate the devastating nature of the disease and this can impact upon how people respond to care.

**Design.** Hermeneutic (interpretive) phenomenology: suitable for studying lifeworld experiences.

**Method.** Life story interviews were conducted with four participants and subjected to interpretive analysis.

**Results.** Three phases of the MND illness trajectory emerged: 'body failing prematurely and searching for answers', 'body deterioration and responses to care', and 'body nearing its end and needing to talk'. These phases highlight the phenomenon under study, all relating to uncertainty for people living with MND.

**Conclusions.** This study showed that people with MND are living with uncertainty and other concerns throughout their illness trajectory. People are having to turn to palliative care professionals who are more able to meet their concerns than those caring for other aspects of their disease.

**Relevance to clinical practice.** MND is a complex disease and it is important that professionals continue to provide holistic care throughout the illness trajectory. The identification of three distinct phases of the MND illness trajectory will help nurses and other professionals to better understand the meaning of uncertainty and other concerns for people with MND.

**Key words.** *Body experiences, concerns, motor neurone disease, palliative care, uncertainty.*

## METHODS

### BACKGROUND

Communication of the diagnosis of motor neurone disease (MND) is 'daunting' for both neurological consultants and individuals (Aoun et al., 2017a, p.368), and anxiety before a diagnosis can be increased as a result of experiences during a long period of investigation (Bäumer et al., 2014), with the median time from onset of symptoms to diagnosis being 15.6 months (Donaghy et al., 2008). When a person receives a diagnosis of MND, they learn whether they have the inherited (familial) form of MND, which occurs in only 5-10% of people diagnosed; or the more common sporadic form of MND, where the disease develops through a combination of genetic susceptibility and environmental factors. They also learn that MND is a progressive and terminal disease resulting in the degeneration of motor neurones, which can affect their movement, speech, respiration and swallowing (Bäumer et al., 2014) and may also affect their cognition and/or behaviour (Goldstein and Abrahams, 2013). The rate of progression and symptoms experienced vary and this is dependent upon the type of MND diagnosed. The types of MND include: amyotrophic lateral sclerosis (ALS), progressive bulbar palsy (PBP), progressive muscular atrophy (PMA) or primary lateral sclerosis (PLS) (Talbot et al., 2008). A person diagnosed with PBP MND may experience a rapid decline and die within a few months, whereas an individual diagnosed with ALS MND may experience a slower decline and die three years after symptom onset (Mitchell and Borasio, 2007). MND results in complete dependency on others, especially towards end stage care (Dawson and Kristjanson, 2003), and individuals find themselves having to be involved with many healthcare professionals.

Research into the lived experience of MND highlights feelings of uncertainty before diagnosis because of fear of the unknown (Ozane and Graneheim, 2017). Receiving a diagnosis of MND can be shocking (Brown, 2003) and be perceived as a death sentence (Locock et al., 2009). People experience the loss of motor ability early into their illness trajectory and this impacts on their occupational roles (Brott et al., 2007), resulting in their experiencing MND as a disruption to their lives (Locock et al., 2009) and having concerns regarding their losses and their future (Brown and Addington-Hall, 2008; Foley et al., 2012), and feelings of uncertainty throughout their illness trajectory because of not knowing when or how they will die (Harris, 2015). Family carers report that the long illness trajectory of MND can create existential suffering for a person with MND (O'Toole, 2011) and an excessive burden for their family carer(s), which coupled together may exacerbate a 'patient's desire to hasten their death' (Whitehead et al., 2011, p.368). The lived experiences of MND accentuate the devastating nature of the disease and this can impact upon how people respond to care.

MND is recognised as a complex and complicated disease (Hogden et al., 2017; MNDA, 2015), and it results in people having to be involved with MND professionals who are disease specialists (Ozanne et al., 2013). Hogden et al. (2017, p.210) suggest people with MND are 'often required to make decisions for symptom management and end-of-life care. Symptom management includes: gastrostomy, invasive ventilation and non-invasive ventilation, to provide improvements in quality of life, as the disease progresses'. Gale (2015, p.251) suggests thinking about the future with further losses and planning for end-of-life is 'serving the agenda of the health professional at the expense of the patient'. Studies of the lived experiences of MND care have suggested that people were generally dissatisfied with the focus of their care (Bolmsjö, 2001; Brown, 2003, 2005; Hughes et al., 2005; van Teijlingen et al., 2001). Although professionals acknowledge the debilitating impact of MND on a person, the focus of care is functional rather than emotional (Brown, 2003). The call is for professionals to change their priorities and to carefully listen to individual accounts of their lived experiences, thus treating the person as an 'individual rather than a patient' (Bolmsjö, 2001, p.503). Miles and Asbridge (2014, p.286) also acknowledge the need for modern medicine to respect the whole person, stating: 'the patient is a person with dimensions which extend well beyond the purely physical and which include the psychological, emotional, existential/spiritual and social components of human existence'. Ozanne et al. (2013, 2017) propose a solution through respecting the whole person with MND, and suggest the need to involve MND professionals who are specialists in the disease together with specialists in palliative care. Connolly et al. (2015) also recommend the integration of palliation for people with MND to alleviate distressing physical, psychosocial and existential concerns. Hensch and Danielson, (2009, p.228) explored the existential concerns of people with cancer, which they classified into the two main themes of the 'struggle to maintain self-identity' and 'threats to self-identity'. Some of the components relevant to 'threats to self-identity' included 'bodily changes, uncertainty, loss of meaning, loss of control, loss of relationships, and fear of dying'. There are parallels to be drawn for people with cancer and people with MND, as they are diagnosed with a terminal illness/disease, resulting in existential concerns (Bolmsjö, 2000, 2001). The aim of this study was to explore the meaning of living with uncertainty for people diagnosed with motor neurone disease (MND).

## Design

The current research is part of a hermeneutic phenomenological project concerned with the meaning of living with MND. The initial study undertaken to answer the research question: '*What does it mean to be a person living through the illness trajectory of MND?*' and a study of the phenomenon of existence when given a diagnosis of MND in the context of receiving healthcare.

The concept of 'existential loss' identified in relation to MND was the loss of past ways of being-in-the-world, and the loss of embodiment, spatiality, and the future (Harris, 2015, p.1579). From the data it emerged that people with MND are living with uncertainty throughout their illness trajectory, and it was determined that this required further examination. To meet the aim of the current research, hermeneutic (interpretive) phenomenology was used as a methodology, as this is suitable for studying lifeworld experiences (Finlay, 2011). Finlay (2011, p.89) suggests 'both descriptive and hermeneutic (interpretive) designs may be used to explore how everyday experience shows itself in the lifeworld, i.e. as embodied and lived through time/space and in relationships with others'. Descriptive phenomenology as a methodology offers researchers an approach through which to explore body experiences and a researcher 'brackets' their existing presuppositions (experiences and beliefs) of a phenomenon in order to promote validity (Husserl, 1989 cited in Allen-Collinson and Pavey, 2014). Hermeneutic (interpretive) phenomenology as a methodology offers an approach through which to explore the body experience and how this impacts upon everyday lives, and a researcher brings their prior presuppositions of a phenomenon to the 'hermeneutic circle' of understanding (Finlay, 2011; Heidegger, 1927/1962). Thus, they are open to a new and evolving understanding of something, such as a 'text', 'phenomenon' or 'participant' in the research context (Finlay, 2011; Heidegger, 1927/1962; Gadamer, 1975).

## Participants

Four people with MND were recruited from the MNDA Care Centre in the North West of England, with the inclusion criteria that individuals had an accepted diagnosis of MND (all types), over 18 years of age (no upper limit), had been receiving treatment from a range of professionals, and had been receiving care and treatment for three to six months. Individuals were excluded from the study if they did not have the mental capacity to provide their informed consent. An MND consultant and nurses provided the study information sheet to people attending the clinic, and asked that they contact the researcher (DH) if they wished to discuss and be involved in the study (Harris, 2014). The participants contacted the researcher to express their wish to be involved and interviews were conducted and at a time and location convenient to them, either in their home or at their place of work.

## Data Collection

Data was collected in 2012. In the initial study the researcher (DH) asked: "Can you please tell me the story of your life.... since you first thought there might have been something wrong with you? Please begin wherever you like. I will not interrupt you; I will listen to you and only write down some

notes. When you finish I may ask you some questions to clarify some points". The researcher (DH) returned to the themes described in the first stage of the interview to explore and gain a deeper understanding of the participants' lived experiences. Data from the first interview was audio-taped and analysed before carrying out the second interview. All participants attended a second interview, which was again at an agreed date/time, when a summary of their lived experiences was discussed, thereby providing an opportunity to add or remove any detail(s). The researcher then asked three open ended questions to induce further discussion.

## Analysis

Interpretive description guided the analysis (Finlay, 2011), and this involves the concept of returning to the whole-parts-whole to facilitate the process of understanding (Heidegger 1927/1962; Gadamer, 1975; Fleming et al., 2003). Fleming et al., (2003, p.118) suggest 'gaining understanding of the whole text should be the starting point of analysis, because the meaning of the whole will influence understanding of every other part of the text'. The current analysis process involved:

1. Returning to the individual philosophical framework analysis.
2. The identification and extraction of excerpts that represent a participant's description of the everyday experiences and how this leads to uncertainty for people diagnosed with MND.
3. Producing a description to reinforce the initial interpretation.
4. Going back and forth between participant descriptions, initial interpretations, themes and forming patterns of the coherent whole.
5. Noting similarities and differences across all the individual excerpts.
6. Grouping together the experiences of the 'body failing prematurely and searching for answers', 'body deterioration and responses to care', and 'body nearing its end and needing to talk'; these illuminate the patterns that represent the phenomenon under study (Finlay, 2011): the meaning of uncertainty for people with MND.

## Plausability of Interpretations

To promote a level of self-awareness, the researcher (DH) reflected on her prior understandings of MND and experiences of supporting her mother living through the illness trajectory of PBP MND, before data collection commenced, and those evolving during the study. To ensure that interpretive closure was not reached prematurely, the interpreted stories were taken back to the participants, and the researcher held regular discussions with other professionals acting as supervisors for the initial study (Harris, 2014). To ensure the researcher's (DH) interpretations are both plausible and

justified (Finlay, 2011), extensive excerpts are provided. The knowledge claims have been validated in dialogue with other professionals who are co-editors of this paper (Finlay, 2011).

### **Ethical Considerations**

Permission to undertake the study was obtained from the researcher's University Research Ethics Panel, the NHS Health Research Authority, and the local NHS Research and Development (approval No. 11/EE/0364). The information sheet and consent form sought approval for participation in the study, primary and secondary analysis, and related publications. Pseudonyms have been used throughout when referring to the participants involved in the study to ensure anonymity. The criterion of avoiding harm was a basic ethical principle in the design of this study.

Hollway and Jefferson (2000, p.87) state 'Can we assume that it is necessarily harmful to experience being upset or distressed? It can be reassuring and therapeutic to talk about an upsetting event in a safe context'. If participants started to get upset I gave them time to express their emotions, and made them aware of support services available to them.

### **Results**

The results of this study highlight three aspects of the illness trajectory of MND: 'body failing prematurely and searching for answers', 'body deterioration and responses to care', and 'body nearing its end and needing to talk'. These bring to light the patterns that represent the phenomenon under study (Finlay, 2011), the meaning of uncertainty for people with MND. Other existential concerns, including bodily changes, loss of meaning, loss of control, loss of relationships, and fear of dying and death, can be found throughout their stories.

### **Body failing prematurely and searching for answers**

The body failing prematurely and searching for answers from neurological professionals. The similarities drawn, the participants are living with uncertainty of what condition they may have. Thereafter, their experiences of investigations and diagnosis were varied. James, a professional sportsman, took himself back to the significant sporting events occurring at that time and being uncertain as to why he needed to be admitted into hospital for investigations:

*So on the last day of the Ryder Cup, I remember it well, I found myself being admitted to [Name of hospital] neuro ward for more electro-nerve tests. And then on the following day I think I had a lumbar puncture, and then I had a brain scan, MRI brain scan, and was then diagnosed with MND.*

*I could probably have done them all in a day, or a day and a half. But obviously it had to fit in with all the test equipment, and the time involved, and the scheduling of the department.*

*When I was in [Name of hospital] being diagnosed there was an awful lot of people came in to look at me, because I think I have quite physical fasciculations, which are a diagnostic indicator of things not going well.*

Martin first noticed something untoward was happening to his body when he dropped his pencil whilst drawing. His GP recommended he went private for neurological investigations and consultancy as changes to his body had a profound impact on his role as a managing director/architect:

*The first time I suspected there was something wrong, was I am or I was left handed, whereas now I am not any handed of course, but I was left handed. But I started dropping things and being clumsy with my right hand, um I knew it was out of character; actually I have been blessed with a lot of skill, in being able to do anything mechanical with my hands for drawing.*

*So I arranged for a private consultation with (name of MND consultant) at the (name of hospital), which I got one in 2/3 days. He then arranged for an MRI scan on the Friday evening and the results then to be analysed on the Monday morning.*

Charles was out walking his dog when he first noticed his foot drop. His physiotherapist referred him for private investigations and consultancy. The consultant's approach to diagnosis was potentially insensitive, as the period after was experienced as life in crisis because of Charles' loss of future relationships with his family:

*I had taken early retirement, at quite an early age, at 53, and therefore I enjoyed some years of retirement before diagnosis. So therefore can be a little bit more philosophical about life, than had it hit me when working, and suddenly life had to change all of a sudden, to give up work, and to give up that particular income stream, and for my whole life to change. So that probably did make it easier to er... cope with it.*

*Rightly or wrongly I was still covered by some private health that did make a difference, that was Thursday and I was expecting an answer by Monday morning.*

*The guy said 'You have been told on your own, go away and talk to your wife, and come back next week or ten days later when you have looked at the internet and found out everything.' He said 'Don't look at the worst bits, because you know what the internet is like, it always gives you the worst things.' He said 'You know it is not good but come back and...' He said 'We will book a double session', whatever that was, so we just went back... [Wife] and I to talk to the consultant in his rooms in a very relaxed way, and giving every single question you can think of.*

*When news suddenly comes out of the blue and it very much was so, er... The period after that you are in tatters really, both yourself and your immediate family.*

*But even my own feelings have changed from initial thoughts of Dignitas type routes to how you want to end it in the end.*

Maureen endured many months of investigations involving a number of hospital admissions, and she experienced a loss of control during the process of investigations and diagnosis, resulting in uncertainty regarding what it all meant:



*I had to go home and then come back in about a month later. Then I went in and then they did a lot of tests, like all the nerve ends of your body, and all that, and a lot of other tests. You know I can't just remember all of them, but I had a lot of tests. I had a full scan and then I went back, and then they said we'll have you in for five days. Anyway they did all tests on me and that five days lasted for about eight weeks.*

*And then I had all the students around me seeing, because you've got to let people learn, haven't you? I don't mind that.*

*And when (name of MND consultant) said 'Do you want the good news or the bad news?' I said 'I will have the bad news first'. He said 'You've got what they call motor neurone, do you know about it?' I said, 'No, I've never heard of it'. And then he said 'You can lose, you can lose your speech, your walking', he said you can lose everything except your brain. I never asked about your eyes, but he said you can lose everything but your brain; that keeps going.*

## Body deterioration and responses to care

Body deterioration and responses to care from MND professionals vary depending on whether a person accepts life with MND. The similarities are however, that the participants are living with uncertainty as to how their care will be managed:

*But you know they keep on talking about managing my symptoms. I don't suffer any pain, so I'm not totally certain what symptoms we have that need managing. (James)*

*I am supposed to be under (name of head MND consultant) at (name of hospital), I have never seen (name of head MND consultant) ever, and the doctor that I usually see is (name of second MND consultant). (Martin)*

*Now when I go to the MND centre I don't spend very long with those individuals because I have really got everything I need locally. So the teams have, NHS related professionals have been excellent. (Charles)*

The progressive nature of MND means involvement with a Multi-professional team of professionals who carry out their individual assessments of need, and this approach was experienced as enlightening, overpowering, and annoying:

*Well I suppose it's a bit of shock being in a situation where you have to be on the receiving end of quite a lot of healthcare. It suddenly makes you wake up to the... either the supreme efficiencies, or deficiencies of the NHS as a healthcare provider. And that er... that definitely has raised a few eyebrows. (James)*

*Receiving healthcare, some people find it overpowering we have had someone visit every day this week, if you include yourself as a health professional. Take yesterday, the district nurses rang, we have had the speech and language therapist, and the lady from the MND association come to see me. I have been to the hospice on Monday. (Charles)*

*I had the speech therapist, and she assesses your food and how you eat, and what I eat. She came about a fortnight ago. I very rarely see a social worker, and I am a bit annoyed with them. I don't want to see them. I have the nurse and my physiotherapist. I have a new OT, I am*

*just beginning to get used to, but I have to keep mithering and mithering for things. I don't want me home being like hospital, I definitely don't. (Maureen)*

The progressive nature of MND means people have no choice but to accept care from formal carers for their survival, although how they respond to this relationality is different for each person. The formal care received via social services was essential for attending to basic human needs; however, James raised concerns regarding threats to maintaining his self-identity, such as managing his daily insulin, as he will be dependent on district nurses to administer it, thus adding yet another professional coming to his home each day, he stated:

*The problem is, you see, you have got social services looking after the personal care side of things, which they are doing, and then you've got the medical practitioner... You know, when I get to a stage, should I ever get there, that I'm not in a position to self-administer my insulin then the care people can't do that, somebody else has to come in and take care of that, which considering what is involved strikes me as a real sledgehammer to crack a nut.*

Maureen, a retired carer, had a prior understanding of the provision of day care. She describes a loss of relationship with a carer, who was like a daughter, coupled with loss of control of self in her world, adds to her frustration:

*I think it was when I worked on [Home Help], some of them didn't have families, I used to feel sorry for them, and I used to go in my own time, you know, at night, or back if they were on their own.*

*My day care is getting better, because like when my first carer left, she went to Australia, she was like my daughter. She always said I was like a second mother. And she did care. But now they just come and do, and feed you, and all that.*

*But now I am getting that way, that if there's a few in and they are talking, I feel as though I am getting worked up. And I am getting more worked up when I have to phone people over complaints and that, I am getting worked up very much, but I was never like that.*

Maureen had experienced a number of hospital admissions, resulting in her spending long periods of time in an acute setting, and having to make decisions on essential medical interventions, such as non-invasive breathing equipment and overnight carers, as without them she could not survive. Thus Maureen is struggling between needs and wants; she knows that she needs the care but does not want the care.

*I was in [name of respiratory hospital] for a long time, they wouldn't let me come home until I decided that I'd have to have night care, and I didn't want it. I didn't want it.*

*I'm on a ventilator overnight, but I've had the mask over my nose, but now I'd rather have the things in my nose because I can breathe better and I don't feel closed in. I have night carers now.*

*Anyway I had to decide then I'd have it because... everything went wrong in my body, you know, everything went wrong.*

*And that is why they are here, because if it comes out they have to be here to put it back, or if I choke two or three times where I have to have it off a bit to get right to sit up, because I feel as though I am choking.*

*I never wanted them [overnight carers], but I would be lost without them.*

## Body nearing its end and needing to talk

The body nearing its end and needing to talk with palliative care professionals occurs in the second to tenth year of illness trajectory. Similarities can be drawn, in that all the participants are living with uncertainty as to how they will be supported to die with MND. James prioritises his need to talk because of a loss of meaning and frustration as he comes to terms with dying:

*Erm... I am getting on with life it's the only way to go. And of course that's being amply demonstrated by the fact that the football season has started, Grand Prix season has started, football season is boiling up to a fantastic climax, and you just think to yourself, well, hm, am I going to see the end of the football season? Or am I going to see the.. Who knows? This is a tricky one.*

*But the snag is, as I think I mentioned earlier, in this instance, maybe my positive mental attitude has been beaten by the fact that I can't stop this disease, this degeneration from continuing. And maybe that's not a good basis on building a solid positive mental attitude. It's a bit like building a house on quicksand, it doesn't matter how solid the building is, and if you haven't got a decent foundation we're never going to succeed. And I suppose, if I'm honest, I am probably phlegmatically resigned to my fate, rather than fighting it from every corner. But that's... you know, that's with the wisdom of two years hindsight.*

*It's a funny old situation. You sort of... it's like being in... It's like being on death row without knowing when the executioner is going to pop in. Anyway, we shall be sorting that one out.*

*Well, funnily enough, one of the things that erm [Name of consultant], at my last clinical appointment, put me... gave me a referral to [Name of hospice consultant], I think he's in palliative care at the hospice, and has a clinic at [Name of hospital], but bearing in mind it's April, I haven't heard a dicky bird since. If they don't get on with it I'll be a name on a list of deceased patients, rather than somebody that is on a palliative care list.*

James had concerns for his wife and children after his death, and searched for counselling services who would understand about their loss, he self-referred to the hospice Macmillan nurse:

*I did go to my GP surgery to find out about counselling services, or what was available for my wife, and my children, after my death, and the first foray into that wasn't hugely successful. I think if you weren't a single mum with an alcohol narcotic problem it was quite difficult to put somebody in the right pigeon hole, but eventually I found them myself, through the specialist Macmillan nurse.*

*And more importantly to me, which I thought was probably significant, she's obviously working at the hospice, so she has had quite a number of years in dealing with patients with a terminal illness, which obviously makes my visits to her, as far as I'm concerned, a lot more grounded than they would be with a counsellor that just deals in general counselling.*

Martin raised his concerns that MND professionals who specialised in the disease are not open to having discussions about the terminal stage of the disease:

*I mean I think people... Because it is a terminal disease are always a bit frightened, they have to have somebody with them at first, they are frightened of being politically incorrect. Which is... You know they are frightened of ticking the wrong box. Of course he doesn't bother now, I... I'd rather he speak frank and open.*

*I mean so many people, and I find it probably a little bit disappointing with some of the staff from the occupational therapy, they are frightened to mention the word 'terminal'.*

Martin appreciates not needing to forward plan an appointment to talk to his hospice consultant about his condition; although he notes the problems of other professionals not wanting to talk about dying:

*Well, really there is not much I can do for you, you know he said 'but I would be better off passing you over to the hospice'.*

*But I have got nothing but praise for the hospice in every respect. I suppose it's a bit of a problem for doctors, um they are used to being able to find a cure for somebody, but (name of consultant) at the hospice had been very good, and rather than make an appointment I just give him a call when I want to see him.*

*I mean a lot of people don't want to talk about it, but it certainly doesn't worry me talking about it. Well we are all going to die, and I think it will come to every one of us. I suppose we have just been told it is going to... And it is a rather painful death, so...*

Charles focuses on talking to the hospice professionals about his loss of future with his family, future support, and making plans for the future:

*There is actually a local Macmillan nurse who has got expertise in MND and she has come, she is a lovely lady and she is going to be great for me as time goes by.*

*So in the end certainly we are in the midst of putting together the... the/ Living Will type thing, personal directive.*

*But what I do feel is incredibly sad that I won't be around to see things ... and to enjoy my retirement with my family.*

Maureen only goes out of her home once a week to hospice day care, where she is talking about completing an advance care plan, knowing that her religious faith will help her to face the fear of dying and death:

*Because now they give you books to write everything down. But you have got to accept things haven't you. I always say well my faith helps me and things like that, you know.*

## DISCUSSION

The results show that people with MND go through three significant body experiences impacting on their everyday lives: body failing prematurely and searching for answers, body deterioration and responding to care, and body nearing its end and needing to talk. Lerum et al. (2015) also raise awareness of three phases of MND: 1. diagnosis, 2. ongoing change and decline, and 3. chronic or terminal. The results of this study focus on categorising MND as a terminal disease from diagnosis, because people with MND are living with uncertainty and other existential concerns relating to their bodily changes, loss of meaning, loss of control, loss of relationships, and dying and death.

During the first phase of the illness trajectory of MND, the theme *body failing prematurely and searching for answers* indicated that a quick diagnosis can lead to living with the uncertainty of what it all means, whilst a longer wait for diagnosis means living with the uncertainty of what condition an individual may have. Consequently, both quick and slow diagnoses can lead to uncertainty of different kinds. One of the issues that emerges from these results is that people with MND are at risk of experiencing life as an existential crisis following a quick diagnosis, and one participant described this period as *"life in tatters"*, relating to difficulty with a loss of future with family, and consideration of a Dignitas approach. Similarly, other studies have shown that people are living with uncertainty before diagnosis and living with a fear of the unknown after diagnosis (Ozanne and Graneheim, 2017). Fear of the unknown may arise because a person newly diagnosed with MND learns that they have a progressive and terminal disease which will lead to the loss of movement, speech, breathing and swallowing (Bäumer et al., 2014). Other study results suggest that a prolonged time to diagnosis and the way a diagnosis is delivered, can result in adverse consequences (O'Brien et al., 2011), such as long-term emotional stress (Aoun et al., 2017b). Emotional stress at the time of diagnosis and the period afterwards has been described as 'shock, fear of the future, denial, anxiety for family, loss of future, anger, isolation, sadness, and guilt' (MNDA, 2013, p.23). A study by Aoun et al. (2017a) suggests that neurologists need to improve their skills relating to responding to the emotions of individuals newly diagnosed with MND. The results of the present study suggest that people experience their body failing prematurely before being diagnosed with MND, and they have a need to talk to a neurological consultant about their concerns of uncertainty and loss of future.

In the second phase of the MND illness trajectory, responses grouped under the theme *body deterioration and responses to care* are varied, depending on whether body deterioration is rapid or slow, and people are living with the uncertainty of 'managing' their condition. Cohn (1997, p.13) states that a 'person has the freedom to accept or deny the conditions of their existence' and also 'a person can choose how to respond to being with others such as choosing involvement or isolation.' The study results suggest that accepting life with MND results in the continuity of life, but not as before, until a person can no longer conceal the fact that their body has deteriorated beyond recognition. Choosing involvement as a response to care meant that individuals had their symptoms monitored re nutrition/hydration and respiratory distress, and their needs were assessed for essential social and medical care, which potentially resulted in a loss of the control of self. Hogden et al. (2017, p.210) suggest 'the consequences of avoiding decision making, for example, for nutrition/hydration or respiratory distress, can impact the patient's health and quality of life.' Other studies suggest people 'engaged with services on their own terms to be in control of care but equated living with ALS to a life of unremitting loss which included loss of control' (Foley et al., 2014, p.118). The study results reveal that people are at risk of experiencing life as an existential crisis approximately two years into their illness trajectory, and one participant described life as "*being a prisoner on death row*", because the present and future were uncertain. Other researchers have reported that physical losses associated with MND place a person's existence on hold (Ozanne et al., 2013). The desire to hasten death has been associated with a loss of control (Bolmsjö, 2001), being unable to accept the disease (Cipolleta et al., 2017), and hopelessness and fear of becoming a burden (Whitehead et al., 2011). This study has revealed that people experience their body deteriorating and have a desire to talk to MND professionals about their concerns of uncertainty, loss of control, and loss of meaning; however, instead they are being referred on to palliative care professionals for this aspect of their care.

The third phase of the illness trajectory, encompassed by the theme *body nearing its end and needing to talk*, refers to the involvement of palliative care professionals because of living with the uncertainty of how they will be supported to die. Participants were referred to the palliative care professionals in their second to tenth year of illness trajectory. Palliative care professionals understand that the phenomenon of dying involves the affirmation of life and regard dying as a normal process (WHO, 2002). Sigrist (2009) suggests the affirmation of life involves finding meaning in life, reflecting on past and present relationships and completing unfinished business. The MND Association (2016) recommend professionals have open discussions about the progressive nature of MND and dying and death to enable people to exercise choice and remain in control of their preferences for end-of-life care. Connolly et al (2015, p435) suggest that early and open discussions

of end-of-life issues may 'obviate the introduction of unwanted interventions and use of technologies'. The participants did not talk about having such early and open discussions of end-of-life issues, although two out of the four participants were using technologies to help them survive. One participant raised his concerns about attempting to have open discussions with MND professionals and stated: *they are frightened to mention the word 'terminal'*. Other study results suggest that, people with MND experience anxiety over dying and death, and recommend referral to palliative care professionals at the time of diagnosis (Ozanne et al., 2013). Goldstein and Abrahams (2013) note that a diagnosis of MND prioritises the need to talk to palliative care professionals early in the illness trajectory because people are at risk of cognitive decline and behavioural changes. Cognitive decline impacts on the ability to communicate advance decisions to refuse treatments (ADRT) or the withdrawal of treatments (NCPC, 2011; MNDA, 2016). This study raises awareness of how people experience their body nearing its end and the need to talk to palliative care professionals about their concerns of uncertainty and fear of dying and death. Talking to palliative care professionals may also enable people with MND to complete unfinished business, find meaning in life, and reflect on past and present relationships with others.

## CONCLUSIONS

The aim of this present study was to return to the everyday experiences of people living with MND and how this leads to uncertainty for people diagnosed with MND. The findings show that people with MND are living with uncertainty and other concerns throughout their illness trajectories. The effects of this can result in people experiencing life in crisis at different stages of their illness, resulting in the desire to hasten death because of various losses. People are having to turn to palliative care professionals who are more able to meet their concerns than those caring for other aspects of their condition. MND is a complex disease and it is important that professionals continue to provide holistic care throughout the illness trajectory. The identification of three distinct phases of the MND illness trajectory will help nurses and other professionals to have a better understanding of the meaning of uncertainty and other concerns for people with MND. The uncertainty that surrounds all phases of MND tends to be overlooked by those caring for people with MND. This research extends our knowledge of existential concerns and how people are being referred to professionals who are specialists in palliative and end-of-life care too late into their illness trajectory, when additional risks associated with a loss of mental capacity and loss of speech may impact on their ability to talk about their concerns and plan for their future.

**Word count:**  
**Abstract 290**





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## WHAT DOES THIS PAPER CONTRIBUTE TO THE WIDER GLOBAL CLINICAL COMMUNITY

The results of this study have a number of important implications for future practice:

- MND is a complex disease and it is important that professionals continue to provide holistic care throughout the illness trajectory.
- The identification of three distinct phases of the MND illness trajectory will help nurses and other professionals to better understand the meaning of uncertainty and other concerns for people with MND. This may help to avoid palliative care patients in general experiencing life as a crisis and having to seek support from hospice professionals too late into their illness trajectory.
- These results further support the concept of providing education for nurses and other professionals on improving the dialogue relating to concerns, in particular uncertainty, and the need for early referral to palliative care professionals.